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CASE REPORT

Successful Weaning of a Laryngeal Mask Airway After a Tongue-lip Adhesion Operation in a Case With Cerebro-costo-mandibular Syndrome

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Cerebro-costo-mandibular syndrome (CCMS) consists of severe micrognathia, glossoptosis, posterior rib-gap defects and developmental delay. It may cause upper airway obstruction and flail chest, resulting in neonatal hypoxia, and possibly death. Early airway management or surgical intervention to maintain a patent airway is critical to avoid hypoxia in CCMS patients. We report a newborn with CCMS who was successfully weaned from a laryngeal mask after undergoing a tongue–lip adhesion operation at 164 days of age.

1. Introduction

Cerebro-costo-mandibular syndrome (CCMS) is a rare congenital disorder of unknown etiology characterized by micrognathia, cleft palate, posterior rib-gap defects and developmental delay.[1,2] Since its first description in the 1960s, more than 50 cases of CCMS have been reported in the literature worldwide.[2] None of the cases have been reported with a chromosome anomaly. Most cases are sporadic, but familial occurrence and an autosomal recessive pattern of inheritance have been reported.[3,4]

Glossoptosis, an upper airway obstruction secondary to micrognathia, may cause neonatal hypoxia and feeding difficulties, resulting in mental retardation with intellectual impairment and failure to thrive.[5–7] Thus, early management to maintain a patent airway is critical. We report a patient with features consistent with CCMS, who underwent a tongue–lip adhesion (TLA) operation after 164 days of conservative management. The infant was successfully weaned from a laryngeal mask airway after undergoing the TLA operation.

2. Case Report

A male infant was born at a regional hospital at gestational age of 37 weeks with a birth weight of 2250 g. The Apgar scores after delivery were seven

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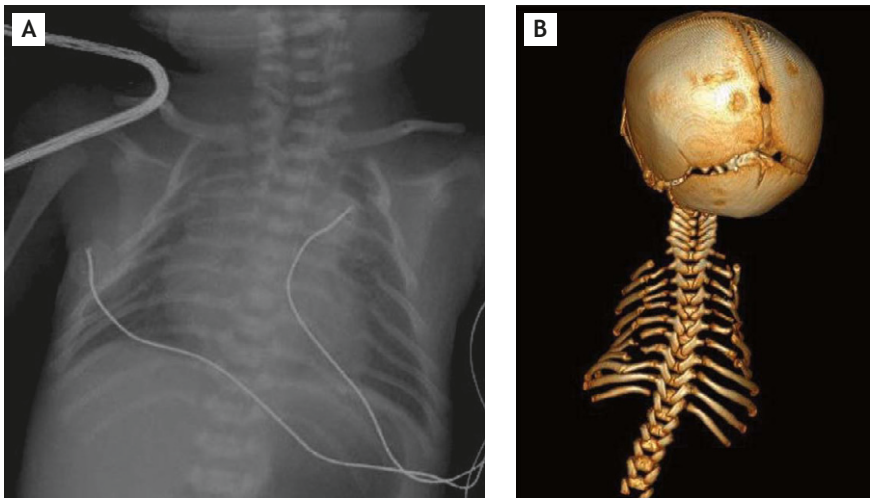

Figure 1 (A) Chest radiograph and (B) three-dimensional computed tomography showed multiple posterior rib-gap defects with flail chest and a bell-shaped thoracic cage.

at 1 minute and eight at 5 minutes. Because of shortness of breath with poor inspiratory movements and generalized cyanosis, he was transferred to our neonatal intensive care unit for further evaluation and treatment. At the neonatal intensive care unit, an attempt to tracheally intubate the patient for frequent desaturation was unsuccessful because of micrognathia and glossoptosis, making it impossible to visualize his larynx. A neonatal laryngeal mask airway (LMA) was inserted temporarily to keep the airway patent.

Chest radiographs and computed tomographic scans showed micrognathia, central cleft soft palate, glossoptosis and multiple posterior rib-gap defects with flail chest (Figure 1). A bell-shaped thoracic cage with multiple rib abnormalities was noted, including the absence of the bilateral 12^{th} ribs and the left 1^{st} rib, gap defects of varying degrees in the right 2^{nd} to 7^{th} posterior ribs and left 2^{nd} to 8^{th} posterior ribs, and a fracture with angulation of the left 9^{th} posterior rib. This presentation was compatible with flail chest. The flail chest compounded the upper airway obstruction and caused respiratory distress. Arterial blood gases showed low O_2 saturation and respiratory acidosis with CO_2 retention, and required frequent insertion of the LMA to assist ventilation and improve saturation. The LMA was intermittently replaced with nasal prongs and continuous positive airway pressure (CPAP) because of the risks associated with prolonged LMA insertion. The maximum duration for each use of LMA was 2 days. In respiratory management of the infant, the LMA was used 20 times and CPAP was used 13 times. The respiratory muscle strength increased as the patient grew, and he was gradually able to tolerate an oxygen hood from several hours to 2–3 days. When considering discharge, we consulted an orthodontic surgeon, who suggested TLA surgery.

The TLA operation was performed at 164 days of age under orotracheal intubation with general anesthesia. The procedure included raising the lower lip mucosa flaps and fixation with the lower edge of the tongue. A solid muscle-to-muscle approximation between the tongue and the lower lip was sutured with three to four 4-0 polydioxanone synthetic absorbable sutures. Finally, two retention sutures with 3-0 nylon were anchored near the tongue base through the lower chin skin surface and tied over silicone buttons (Figure 2). After 1 week of intubation, the patient was successfully weaned from the ventilator. There was marked improvement in respiratory distress without O_2 desaturation and CO_2 retention after operation. In addition, there was no further need for LMA or CPAP. The two retention sutures were removed after 2 weeks when the tongue and lip wounds were healing well. Another TLA was planned for de-adhesion when mandibular growth of the micrognathic mandible had caught up, or the teeth had started to grow.

Feeding difficulties were slow to resolve. Feeding via a nasogastric tube had been the patient's only source of nutrition, and he received daily oral muscle training to encourage the transition to oral feeding prior to surgery. Upper airway obstruction had improved after the TLA operation, but oral feeding difficulty was not resolved. Choking occurred easily, which may have been related to fixing the tongue to the lip and therefore restricting the swallowing movement. As a result, nasogastric feeding remained the only source of nutrition. Body weight and height increased from 2128 g and 49 cm at birth to 3914 g

Figure 2 Photograph of the airway management by tongue–lip adhesion with two silicone buttons.

and 57 cm on operation day 164, and then to 6500 g and 65 cm 3 months after the operation, respectively. The body weight gain velocity increased from 76 g/week before the operation to 201 g/week after the operation. The body height gain velocity increased from 1.5 cm/month before the operation to 4.0 cm/month after the operation. Good social interaction and good limb movements were observed. Continual follow-up of developmental milestones was suggested.

3. Discussion

The treatment protocol for CCMS patients usually starts with conservative management, such as prone positioning, nasopharyngeal airway and LMA. Surgical intervention, such as TLA, distraction osteogenesis or tracheostomy, is considered if conservative management fails. These surgical procedures are used to establish and maintain airway patency.

Prone positioning has been used for glossoptosis by using gravity to displace the tongue anteriorly. Because this is noninvasive and has minimal morbidity, it should be attempted in all glossoptosis patients. High success rates with prone positioning have been reported in a previous study.[8]

Nasopharyngeal airway and LMA placement physically move the tongue base anteriorly out of the airway.[6–8] Both interventions have a higher success rate than the use of prone positioning. However, these interventions cannot be used for long periods of time because the decreased perfusion of pharyngeal mucosa and direct compression of surrounding structures can lead to pharyngolaryngeal morbidity. If long-term airway management is anticipated, surgical management is needed.

TLA is a simple surgical procedure, in which the tongue is sutured to the lower lip, pulling the tongue base forward and relieving airway obstruction.[6–8] Douglas described the use of TLA for the treatment of upper airway obstruction associated with micrognathia in 1946.[9] The majority of these children with unsuccessful nonsurgical management can be successfully treated with TLA, and it should be considered as the first surgical intervention. Although the TLA operation is a minimally invasive procedure without severe or long-term complications, the reported complications of the procedure include wound dehiscence, infection, suture lines cutting through the tongue and lower lip, and injury to the tooth buds and Wharton's duct.[6]

Additional surgical intervention may be needed in patients who do not have catch-up mandibular growth or in whom TLA is not successful. Distraction osteogenesis of the mandible can offer definitive correction of micrognathia.[7,8] McCarthy et al[10] first reported its use on the mandible in 1992. The mandibular angle and body were exposed through bilateral submandibular incisions, a sagittal osteotomy was made in the retromolar region and an internal device was fixed with miniplates and screws. Risks associated with this procedure include injury to the premolar tooth buds, the marginal branch of the facial nerve and the inferior alveolar nerve. Tracheostomy is often considered as a last resort treatment for airway obstruction, particularly because it can cause lasting issues with granulation tissue formation and tracheal stenosis.[8] Tracheostomy can only be used in children with airway obstruction at sites other than the tongue base.

In conclusion, children with CCMS experience upper airway obstruction, which causes respiratory distress and feeding problems. Early management of airway obstruction will improve the growth and development of these children. Our patient underwent TLA after 164 days of conservative management. To our knowledge, this is the first published case report of TLA operation for a child with CCMS. His long-term progress will be closely followed.

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